2017 Revised Classification of Seizures

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The International League Against Epilepsy (ILAE) is the world's main scientific body devoted to the study of epilepsy, and it has recently revised its classification of seizures. The changes will help make diagnosing and classifying seizures easier and more accurate. In this article, you'll find the new general outline of basic seizure classification and a more detailed view of classifying seizures.

Background

What is a seizure classification?
Explaining the new classification system requires some background. People with epilepsy have recurring seizures that often occur spontaneously and without warning. The official definition of a seizure is "a transient occurrence of signs and/or symptoms due to an abnormal, excessive or synchronous neuronal activity in the brain."

- This means that during a seizure, large numbers of brain cells are activated abnormally at the same time. It is like an "electrical storm" in the brain.
- The nature of the seizures depend upon many factors, such as the person's age, the sleep-wake cycle, prior injuries to the brain, genetic tendencies, medications, which circuits in the brain are involved, and many others.

Separating seizures into different types helps guide further testing, treatment, and prognosis or outlook. Using a common language for seizure classification also makes it easier to communicate among clinicians caring for people with epilepsy and doing research on epilepsy. The classification also provides common words for people with epilepsy and the general public to describe seizures.

History of seizure classification:

- For decades, the most common words to describe seizures were grand mal and petit mal. Although the medical meaning of these terms was fairly precise, lay people often used them loosely when referring to any big or little seizure.
- In 1981, a classification was developed that has been used for 35 years. This system divided seizures into partial (focal) onset and generalized onset seizures. The partial seizures were further divided into simple partial seizures (no change in consciousness) and complex partial seizures (impaired consciousness). Generalized seizures were divided into various subcategories.
- This classification served well but had several drawbacks.
    - Several important seizure types were not specifically listed, for example, focal clonic seizures or infantile (epileptic) spasms.
    - It was impossible to classify a seizure if the onset (the part or network of brain involved in generating or starting the seizure) was not known.
    - Many of the terms, such as psychic seizures or complex partial seizure were confusing. Some people with epilepsy felt that there was nothing "simple" about simple partial seizures.
- These concerns led to the ILAE's current revision.

The New Basic Classification

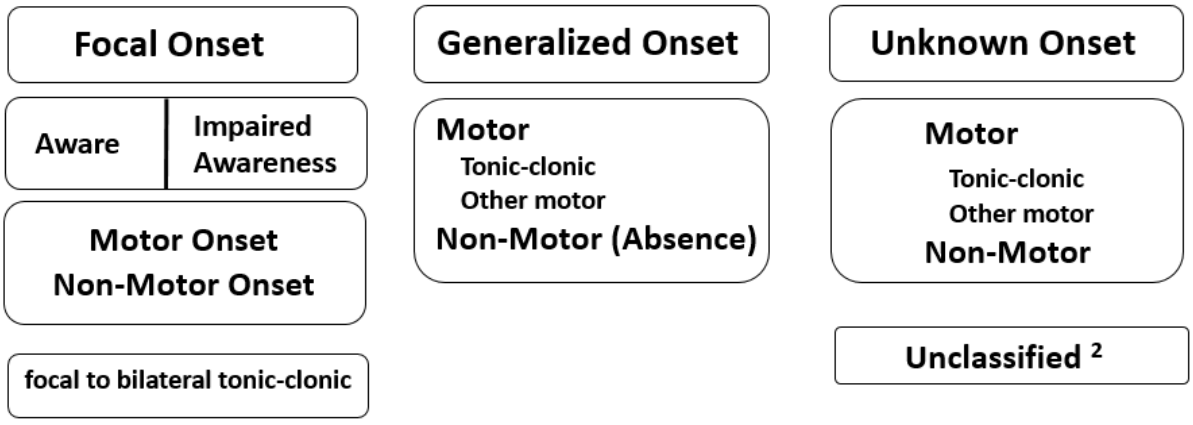
Defining where seizures begin: The basic classification is a simple version of the major categories of seizures. The first step is to separate seizures by how they begin in the brain.

- The classification begins by dividing seizures into those that start focally, meaning involving circuits (networks) in one hemisphere or side of the brain versus those that engage networks in both sides of the brain at the onset.
- If onset is unknown, the seizure falls into the unknown onset category. Later on, the seizure type can be changed if the onset of a person's seizures becomes clear.
- The type of seizure onset is important because it affects choice of seizure medication, possibilities for epilepsy surgery, outlook, and possible causes.

Describing awareness in focal seizures: The level of awareness is of practical importance because it is one of the main factors affecting a person's safety during a seizure. Awareness is a surrogate marker for level of consciousness, but it is simpler to evaluate than the more complex concept of consciousness.

- *Focal Aware* - A seizure is "focal aware" if awareness is intact, even if the person is unable to talk or respond during the seizure. This replaces the term simple partial.
- *Focal Impaired Awareness* - A seizure is classified as "focal impaired awareness" if awareness is impaired at any time during the seizure. This replaces the term complex partial seizure.
- Sometimes it's not possible to know if a person is aware or not, for example if a person lives alone or has seizures only at night. In this situation, the awareness term may not be used.
- A seizure can start focally in the brain and spread to both sides of the brain, resulting in a "focal to bilateral tonic-clonic seizure." This used to be called a secondarily generalized tonic-clonic seizure. The word "generalized" is now only used if seizures are known to affect both sides of the brain at once in the beginning of a seizure.
- The term aura is often used to describe symptoms a person may feel in the beginning of a seizure and is not in the new classification. Clinicians will continue to understand the use of the word aura. It's important to know that in most cases, these early symptoms may be the start of a seizure.

ILAE 2017 Classification of Seizure Types Basic Version [1]



[1] Definitions, other seizure types and descriptors are listed in the accompanying paper & glossary of terms

[2] Due to inadequate information or inability to place in other categories

Describing motor and other symptons in focal seizures: Many other symptoms may occur during a seizure. In this basic system, seizure behaviors are separated into groups that involve movement.

- **A focal motor seizure** means some type of movement occurs during the event. For example, twitching, jerking, or stiffening movements of a body part or automatisms (automatic movements such as licking lips, rubbing hands, walking, or running).
- **A focal non-motor seizure** includes other symptoms such as changes in sensation, emotions, thinking, or experiences.
- A seizure therefore can be focal motor (the word "onset" is implied) or focal non-motor, for example, focal sensory, with or without mention of awareness.
- It is also possible for a focal aware or impaired awareness seizure to be sub-classified as motor or non-motor onset.

Describing generalized onset seizures: Seizures that start in both sides of the brain, called generalized onset, can be motor or non-motor (absence).

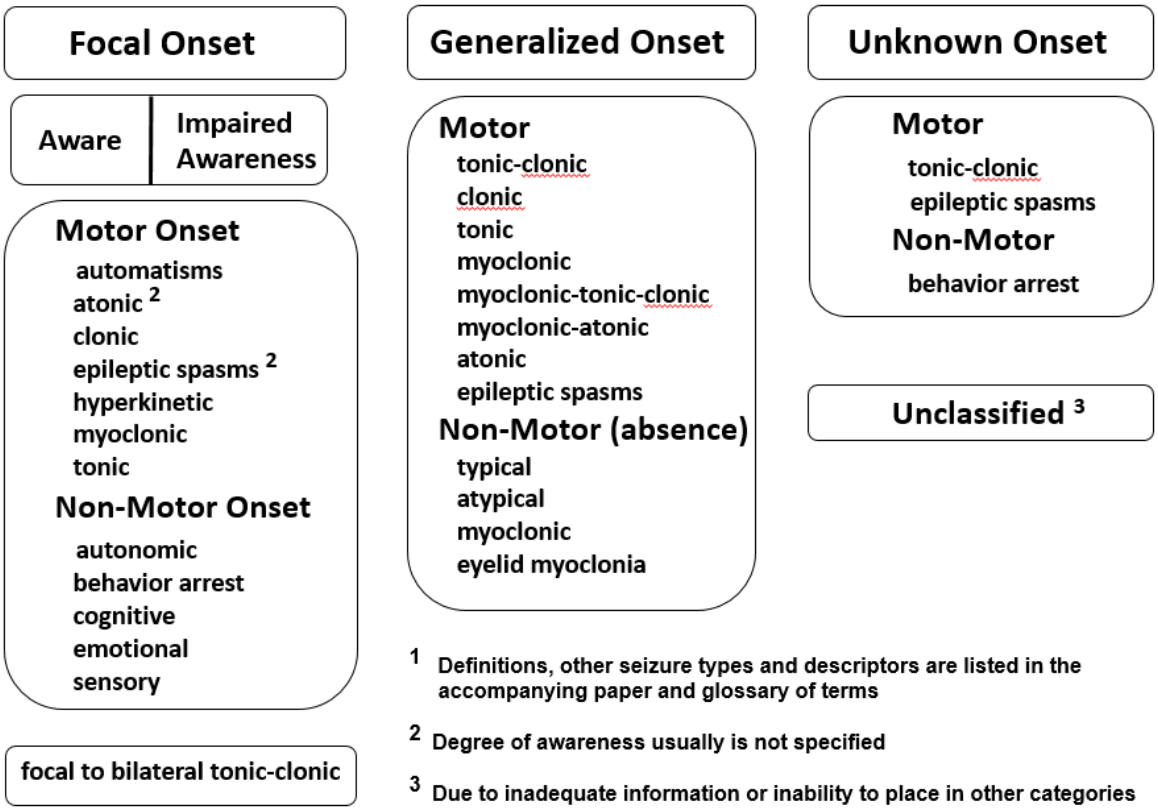
- The generalized tonic-clonic seizure term is still used to describe seizures with stiffening (tonic) and jerking (clonic). This loosely corresponds to "grand mal."
- The generalized absence seizure corresponds to the old term "petit mal." These seizures involve brief changes in awareness, staring, and some may have automatic or repeated movements like lipsmacking.

The New Expanded Classification

The expanded classification keeps the framework of the basic classification, but adds more seizure types as subheadings.

- **Focal motor onset seizure** types include automatisms, atonic, clonic, epileptic spasm, hyperkinetic, myoclonic and tonic seizures. Several of these types also appear in the generalized onset categories. (see below for descriptions)
- **Focal non-motor onset seizures** include autonomic, behavior arrest, cognitive, emotional, and sensory seizures. Since seizures often have several different symptoms and behavioral signs, the seizure is named for the first prominent symptom or sign. This has been the usual clinical practice, because seizure onset marks the part or network of the brain involved in generating the seizure. Other regions become involved as the seizure spreads.

ILAE 2017 Classification of Seizure Types Expanded Version [1]



More Information About Different Seizure Types

Below are brief descriptions of each seizure type.

Focal onset seizures may occur with or without impairment of awareness, except that atonic and epileptic spasm seizures usually do not show obvious impairment of awareness.

- *Focal automatisms seizure*: A seizure with automatic fumbling behavior, such as lip-smacking, hand-rubbing, picking at objects, walking in circles, repeatiting meaningless phrases, or undressing.

- *Focal atonic seizure*: Focal, for example in one arm or leg, sudden loss of muscle tone and strength, resulting in a transiently limp limb.

- *Focal clonic seizure*: Sustained rhythmically jerking of one part of the body or face.

- *Focal epileptic spasms*: Sudden flexion or bending of the trunk with flexion or extension of the limbs lasting less than a few seconds. These often occur in clusters. The term infantile spasms applies to epileptic spasms occurring during infancy. Video-EEG monitoring and a brain MRI may be needed to determine whether onset of epileptic spasms is focal or generalized.

- *Hyperkinetic seizure*: A seizure with vigorous thrashing or pedaling movements. Even though both sides of the body are usually involved with these seizures, the EEG often shows a focal and frontal lobe origin. Some people used to call these hypermotor seizures.

- *Focal myoclonic seizure*: Irregular and brief lightning jerks of limbs or face on one side of the body.

- *Focal tonic seizure*: Stiffening of arm, leg, or neck producing a forced posture during the seizure.

- *Focal autonomic seizure*: A seizure whose primary effect is on autonomic nervous system functions, such as heart rate, blood pressure, sweating, skin color, hair standing on end (piloerection), and gastrointestinal sensations.

- *Focal behavior arrest seizure*: In this seizure type, movement stops, sometimes called a freeze or a pause. Because brief behavior arrest is common and hard to recognize as being abnormal, a seizure should only be classified as a focal behavior arrest seizure if the behavior arrest is the main feature through the entire seizure.

- *Focal cognitive seizure*: This type of seizure refers to impaired cognition (thinking) during a seizure. The impairment might affect language, spatial perception, ability to calculate math, or other cognitive functions. Do not count loss of awareness or memory (unless only memory is impaired) as a focal cognitive seizure, because awareness is used to describe other seizure types.

- *Focal emotional seizure*: This seizure type begins with spontaneous fear, anxiety, or less often joy. There may be involuntary laughing or crying, each of which might or might not be accompanied by a subjective emotion. Gelastic and dacrystic seizures would fit into this group.

- *Focal sensory seizure*: Sensory seizures can consist of tingling or numbness, visual symptoms, sounds, smells, tastes, tilting or spinning sensations (vertigo), and hot-cold feelings.

Generalized onset seizures are not characterized by level of awareness, because awareness is almost always impaired.

- *Generalized tonic-clonic*: Immediate loss of awareness, with stiffening of all limbs (tonic phase), followed by sustained rhythmic jerking of limbs and face (clonic phase). Duration is typically 1 to 3 minutes. The seizure may produce a cry at the start, falling, tongue biting, and incontinence.

- *Generalized clonic*: Rhythmical sustained jerking of limbs and/or head with no tonic stiffening phase. These seizures most often occur in young children.

- *Generalized tonic*: Stiffening of all limbs, without clonic jerking.

- *Generalized myoclonic*: Irregular, unsustained jerking of limbs, face, eyes, or eyelids. The jerking of generalized myoclonus may not always be left-right synchronous, but it occurs on both sides.

- *Generalized myoclonic-tonic-clonic*: This seizure is like a tonic-clonic seizure, but it is preceded by a few myoclonic jerks on both sides of the body. Such seizures are commonly seen in people with the syndrome of juvenile myoclonic epilepsy.

- *Generalized myoclonic-atonic*: This seizure presents with a few myoclonic jerks, followed by a limp drop. These seizures may be seen in children with Doose syndrome.

- *Generalized atonic*: This is an epileptic drop attack, with sudden loss of muscle tone and strength and a fall to the ground or a slump in a chair. Atonic seizures usually last only seconds.

- *Generalized epileptic spasms*: Brief seizures with flexion at the trunk and flexion or extension of the limbs. Video-EEG recording may be required to determine focal versus generalized onset.

- *Generalized typical absence*: Sudden onset when activity stops with a brief pause and staring, sometimes with eye fluttering and head nodding or other automatic behaviors. If it lasts for more than several seconds, awareness and memory are impaired. Recovery is immediate. The EEG during these seizures always shows generalized spike-waves.

- *Generalized atypical absence*: Like typical absence seizures, but may have slower onset and recovery and more pronounced changes in tone. Atypical absence seizures can be difficult to distinguish from focal impaired awareness seizures, but absence seizures usually recover more quickly and the EEG patterns are different.

- *Generalized myoclonic absence*: A seizure with a few jerks and then an absence seizure.

- *Generalized eyelid myoclonia*: Eyelid myoclonia represents jerks of the eyelids and upward deviation of the eyes, often precipitated by closing the eyes or by light. These may be associated with absence seizures in people with Jeavon's syndrome.

Unknown Onset Seizures

- Clinicians using the classification will identify a seizure as focal or generalized onset if there is about an 80% confidence level about the type of onset. This means that there is significant confidence on the seizure onset and type.

- Seizures without enough confidence about onset are labeled of unknown onset. The most important seizures of unknown onset are tonic-clonic, epileptic spasm, and behavior arrest (which could be either a focal impaired awareness or absence seizure).

- If a seizure onset becomes clarified at a later date, the type will change.

General Comments

Classification of a seizure type is only part of the seizure description.

- Use of other descriptive terms or even free text is encouraged. For example, a "focal impaired awareness tonic seizure" might be described as a "focal impaired awareness seizure with tonic right arm stiffening, followed by right arm clonic jerking."

- Most seizures can be classified by accompanying signs and symptoms. However, added information is useful when available, for example, phone videos, EEG, MRI and other brain imaging, blood tests, or gene tests.
- For practical purposes, long descriptive terms are probably not useful for day-to-day life.

The ILAE 2017 seizure classification replaces the 1981 classification that was used for 35 years. Change in terminology is disruptive and can take a lot of work. Adoption happens over time. The effect of the update should be easier classification of all seizure types, greater clarity, and more transparency of terminology to the nonmedical and medical community.

Long Summary: 2017 Revised Classification of Seizures
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